

A Case Report of Posterior Reversible Encephalopathy Syndrome with a Review of Literature

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ABSTRACT Posterior reversible encephalopathy syndrome (PRES) is characterized by typical neurological deficits and magnetic resonance imaging (MRI) findings. A 17-year-old nulliparous woman with 34-week-old pregnancy was diagnosed with preeclampsia and magnesium sulphate infusion was initiated. Despite this medication and an emergency cesarean section, the patient had repeating seizures. Cranial MRI showed hyperintense signal alterations in the cortical and subcortical regions of the right angular gyrus and, thus, a diagnosis of PRES was made. Presenting with headache, altered mental status and severe systolic hypertension in young women, preeclampsia associated PRES can be easily diagnosed by MRI findings. Conservative management with close monitorization of vital and clinical findings and administration of anti-hypertensive and anti-convulsant drugs would provide the reversibility of PRES related radiological changes in most cases. The clinicians dealing with preeclampsia should be aware of this clinical entity and they should appeal to MRI whenever there is a high index of suspicion.

Keywords: Eclampsia; posterior reversible encephalopathy syndrome

Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey et al. in 1996. Being also known as posterior leukoencephalopathy syndrome, hyperperfusion encephalopathy or brain capillary leak syndrome, this is a clinico-neuroradiological entity. That is, PRES is characterized by typical neurological deficits and distinctive magnetic resonance imaging (MRI) findings. These findings refer to hyperintense signal alterations in the cortical and subcortical regions of the parietal and occipital lobes.¹⁻³

Hypertensive diseases such as chronic arterial hypertension, preeclampsia and eclampsia are the major underlying reason for PRES. Collagen vascular diseases, thrombotic thrombocytopenic purpura, hepatic insufficiency, massive blood transfusion, HIV infection, acute intermittent porphyria and organ transplantation may also trigger this syndrome. The use of immunosuppressive drugs and anti-neoplastic agents is also associated with PRES.³⁻⁵

The symptoms of PRES include nausea, vomiting, headaches, convulsions, mental disorder, visual disturbances and cortical blindness. The prognosis of PRES is usually benign. In many cases, clinical symptoms and radiological signs seem to resolve within a period of days to weeks, after the dissolution of the inciting factor. However, there are a few case reports

which report about the emergence of permanent neurological deficits and status epilepticus due to intracranial hemorrhage and ischemic infarcts. Rarely, the increase in intracranial pressure and particularly the involvement of brain stem may cause death.⁶⁻⁸

Pronounced increase in the utilization of MRI and the development of specific MRI techniques such as fluid attenuated inversion recovery (FLAIR) and diffusion weight imaging (DWI) help to identify cortical and subcortical lesions and to clarify the pathophysiology of cerebrovascular alterations.^{5,6} This case report aims to describe a case of PRES occurring in a woman with eclampsia, make a review of related literature and, thus, to contribute to the awareness of the obstetricians about this potentially life-threatening syndrome.

CASE REPORT

A 17-year-old nulliparous woman was admitted to the emergency unit of the study center due to involuntary spasmic movements. It was learnt that she had 34-week-old pregnancy according to her last menstrual period. There was nothing particular in her medical history and her obstetric follow up was uncomplicated. Physical examination revealed mental confusion and pretibial edema in the patient. Her vital findings were temperature: 37.2°C, pulse: 88 beats/min, respiration: 18 breaths/minute, oxygen saturation: 95% and sys-

tolic/diastolic blood pressure: 190/120 mmHg. Obstetric ultrasonography (Logiq P5, GE Medical Systems, USA) showed a single fetus with 33-week-old biometrical measurements and an amniotic fluid index of 40 mm. Her platelet count and transaminases were normal but there was proteinuria of 300 mg/dl in her urinalysis. The patient was immediately diagnosed with preeclampsia and alpha-methyldopa 1500 mg/day, po (Alfamet, İbrahim Etem Ulagay) was administered as an anti-hypertensive treatment. In order to prevent convulsions, magnesium sulphate (Magnesium Sulphate 15%, Biofarma Medical) infusion was also initiated as a loading dose of 4 grams which is followed by 2 grams/hour as a maintenance dose. Despite this medication, the patient had a seizure 10 minutes later and an emergency cesarean section was done to deliver a male baby with a birthweight of 2195 grams, first minute Apgar score of 7 and fifth minute Apgar score of 7. However, the patient had a seizure three hours after cesarean delivery. Although her neurological examination and electroencephalography were normal, computed tomography (Toshiba TSX-101A, Aquillon 16 Slice, Tochigi, Japan) of the cranium was performed (Figure 1). Since hypoattenuation in both frontal, temporal and occipital lobes was observed in computed tomography (CT) scans, a preliminary diagnosis of PRES was made and MRI (Magnetom Symphony Quantum, Siemens Medical Systems, Erlangen, Germany) of the cranium was carried out (Figure

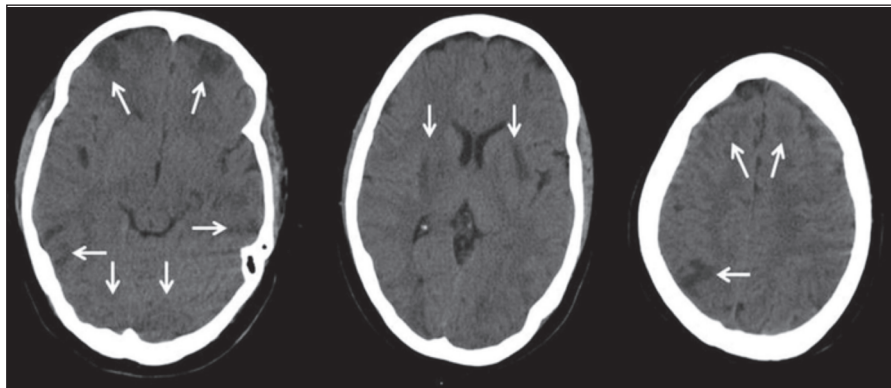


FIGURE 1: Computed tomography scans in axial plane display hypoattenuation in (a) cortical-subcortical areas of both occipital, temporal and especially frontal lobes (b) both external capsules and lentiform nuclei, being more evident in the right lobe and (c) cortical and subcortical areas of the right parietal lobe at the plane of centrum semiovale.

2). The detection of hyperintense signal alterations on the cortical and subcortical regions of the right angular gyrus in coronal T2 FLAIR sequences confirmed the diagnosis of PRES. Anti-hypertensive treatment and anti-convulsant prophylaxis were continued and the patient never had a seizure again. She was discharged from hospital eight days later. Written informed consent was obtained from the patient for publishing this case report.

DISCUSSION

The pathogenesis of PRES remains unclear, but it seems to be related to the impairment in cerebral autoregulation and endothelial functions. The hypothesis that best explains the acute and reversible changes in PRES is that a sudden increase in blood pressure causes dysfunction in cerebral autoregulation and inhibition of cerebral vasoconstriction. The result is extensive cerebral vasodilatation which triggers the extravasation of plasma and appearance of vasogenic edema. Since arteriolar system in posterior cerebrum has weaker sympathetic activity, vasogenic edema becomes more prominent in parietal and occipital lobes.⁸

The pathogenesis of preeclampsia/eclampsia is closely associated with endothelial impairment which prompts the release of angiogenic mediators including endothelin, angiotensin II and thromboxane. These mediators cause vasoconstriction and subsequent cytotoxic edema.⁹

When cases of PRES are analyzed, it can be understood that the number of cases with underlying preeclampsia/eclampsia is significantly higher than the number of cases with other predisposing factors. For instance, Brewer et al. identified PRES as a core component of eclampsia because 98% of the preeclampsia patients had PRES on neuroimaging studies.¹⁰ Roth et al. were the first to indicate that headache was the most frequently encountered symptom in preeclampsia/eclampsia associated PRES.¹¹ They also reported that the patients with preeclampsia/eclampsia associated PRES were significantly younger than the patients with PRES related to other factors. Later, Liman et al. compared 24 women who had preeclampsia/eclampsia associated PRES with 72 patients who had PRES related to other factors.¹² Headache followed by altered mental status were addressed as the most frequently encountered symptoms in preeclampsia/eclampsia associated PRES. Brewer et al. also attested this finding and determined severe systolic hypertension in half of the women with preeclampsia/eclampsia associated PRES.¹⁰ Complying with literature, our case was a young woman who presented with mental confusion and severe systolic hypertension. Raman et al. investigated 92 patients with PRES and found that the most commonly affected region was the parietal and occipital lobes.¹³ However, it was also specified that frontal lobes, temporal lobes, basal ganglia, cerebellum, brainstem and thalamus could be in-

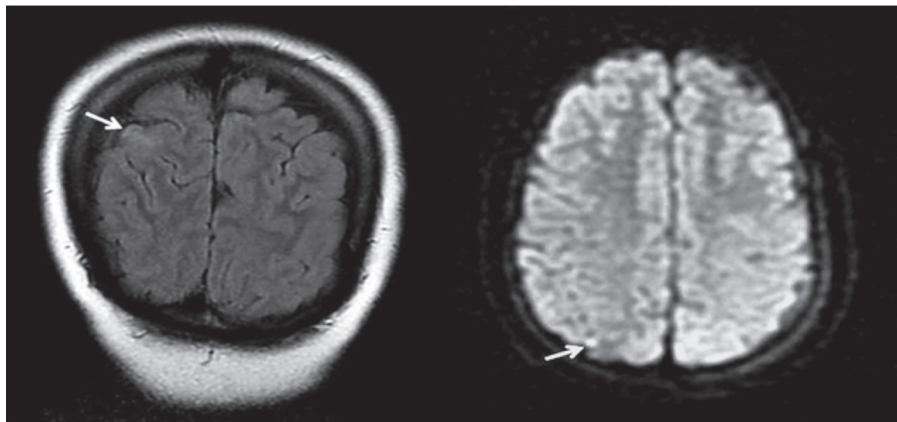


FIGURE 2: Magnetic resonance imaging demonstrates (a) hyperintense signal alterations on the cortical and subcortical regions of the right angular gyrus in coronal T2 FLAIR sequences and (b) mild restricted diffusion on cortical region of right angular gyrus.

volved. Similarly, Fugate et al. who evaluated 120 patients with PRES reported that parietal and occipital lobes were most commonly affected, followed by the frontal lobe, temporal lobe and cerebellum.¹⁴ They also designated an evident involvement of parietal and occipital lobes in preeclampsia/eclampsia associated PRES. In accordance, angular gyrus which is located between parietal and temporal lobes was affected in our case.

Preeclampsia/eclampsia associated PRES was found to have a less severe course than PRES related to other underlying factors. The patients with eclampsia associated PRES had significantly less frequent involvement of thalamus, pons and mid-brain, less edema, less hemorrhage and less contrast enhancement. Moreover, complete resolution of edema was significantly more common and the number of residual structural alterations was significantly lower on their follow up imaging.¹⁰ Pande et al. also concluded that radiological alterations that occur due to eclampsia associated PRES had significantly more reversibility than those that appear due to hypertension and drug related PRES.¹⁵ As for the present case, the patient recovered without any sequelae after conservative management with the sustention of anti-hypertensive treatment and anti-convulsant prophylaxis.

Presenting with headache and altered mental status in young women, preeclampsia/eclampsia associated PRES can be easily diagnosed with the

help of typical MRI findings. Conservative management with close monitorization of vital and clinical findings and administration of anti-hypertensive and anti-convulsant drugs would provide the reversibility of PRES related radiological changes in most cases. The clinicians dealing with preeclampsia/eclampsia should be aware of this clinical entity and they should appeal to MRI whenever there is a high index of suspicion for PRES.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Çiğdem Kunt İşgüder; **Design:** Çiğdem Kunt İşgüder; **Control/Supervision:** Erkan Gökçe; **Data Collection and/or Processing:** Çiğdem Kunt İşgüder; **Analysis and/or Interpretation:** Çiğdem Kunt İşgüder, Mine Kanat Pektaş; **Literature Review:** Çiğdem Kunt İşgüder; **Writing The Article:** Çiğdem Kunt İşgüder; **Critical Review:** Nurşah Başol; **References And Fundings:** Çiğdem Kunt İşgüder; **Materials:** Çiğdem Kunt İşgüder.

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